Medical and Pediatric Oncology 26:145–146 (1996)

Letter to the Editor: The New SIOP (Stockholm) Working Classification of Renal Tumours of Childhood

Based on the correlation between the histological features and survival, three prognostic groups of renal tumours of childhood were discerned in the previous International Society of Paediatric Oncology (SIOP) Nephroblastoma Trials and Studies: favourable, standard, and unfavourable histology tumours [1]. The favourable and standard group together were the same as the favourable group of the National Wilms' Tumor Study (NWTS) Classification [2].

However, during the last decennia a confusion in the nomenclature of subtypes of nephroblastoma developed. To adapt the SIOP nomenclature and classification of renal tumours of childhood to what is presently used in general, the SIOP Panel of Pathologists changed their nomenclature and classification during the meeting (also attended by J.B. Beckwith and A. Kelsey) in Stockholm in June 1994 (Table I). Instead of favourable, standard, and unfavourable histology, the terms "low risk," "intermediate risk," and "high risk" tumours will be used. The histogenesis of the mesoblastic nephroma, clear cell sarcoma of the kidney, and rhabdoid tumour of the kidney is uncertain, but it is believed that they are entities unrelated to nephroblastoma. However, they are all included in the classification since they represent typical renal tumours of childhood. They are printed in italics.

The criteria for recruitment of patients in the SIOP trial and study have been described in the SIOP Nephroblastoma Clinical Trial and Study Protocol 93-01 [3]. According to the protocol, all eligible patients (>6 months or <18 years old at the time of diagnosis, with a unilateral tumour with certain clinical and radiological characteristics of a nephroblastoma, and with no previous antitumour treatment) will receive pre-operative chemotherapy, whereas postoperative treatment depends on the stage defined after surgery and histological subtyping of the tumour. Stage I tumours in the low risk group will recieve no postoperative therapy. The same protocol gives the histological criteria of different subtypes of renal tumours within the three major groups [3]. This new classification is very similar to the original German Paediatric Oncology (GPO) classification (4) except that the latter included only mesoblastic nephroma and cystic partially differentiated nephroblastoma but not other subtypes of nephroblastoma in the low risk group. One certainly might argue whether there is a need for so many histological subtypes of nephroblastoma, but we feel it is essential to identify those renal tumours that would require no postoperative therapy in order not to overtreat the children. Moreover, the results of previous SIOP trials showed that nephroblastoma with fibroadenomatous-like structures [5], nephroblastoma of highly differentiated epithelial type [6], and nephroblastoma completely necrotic after preoperative chemotherapy (preliminary results of the SIOP 9 trial and study, presented by J.F.M. Delemarre at the SIOP Nephroblastoma Committee meeting) had an excellent prognosis and, therefore, have been moved from the intermediate risk tumours to the low risk tumours group.

We believe this classification covers all typical renal tumours of childhood and that it is more straightforward than the previous one. We also hope it will be much easier now to compare our results to those from the NWTS trials in order to find the best treatment for children suffering from these tumours.

SIOP Panel of Pathologists for Renal Tumours of Childhood:

Jan F.M. Delemarre, MD, PhD
Amsterdam, The Netherlands
Bengt Sandstedt, MD, PhD
Stockholm, Sweden
Dieter Harms, MD, PhD
Kiel, Germany
Liliane Boccon-Gibod, MD
Paris, France
Gordan M. Vujanić, MD, PhD
Cardiff, U.K.

REFERENCES

1. Delemarre JFM, Sandstedt B, Gerard Marchant R, Tournade MF: SIOP Nephroblastoma trials and studies, morphological aspects. In Raybaund CI, Clement R, Lebreuil G, Bernard JL (eds): "Pediatric Oncology: Proceedings of the International Society of Paediatric Oncology, Marseille, 15–19 September 1981," Amsterdam: Excerpta Medica, 1982, pp. 261–272.
2. Beckwith JB, Palmer NF: Histopathology and prognosis of Wilms' tumor: Results from the first National Wilms' Tumor Study. Cancer 41:1937–1948, 1978.
3. International Society of Paediatric Oncology (SIOP) Nephroblastoma Clinical Trial and Study Protocol 93-01, June 1993.
4. Schmidt D, Harms D, Leuschner I: Malignant renal tumors of childhood. Path Res Pract 188:1–15, 1992.
5. Delemarre JFM, Sandstedt B, Tournade MF: Nephroblastoma with fibroadenomatous-like structures. Histopathology 8:55–62, 1984.
6. Delemarre JFM, Rey A, Harms D, Sandstedt B, Vujanić G, Tournade MF: The epithelial type of nephroblastoma to be considered as a nephroblastoma with favourable histology? Med Pediatr Oncol 20:435 (A), 1992.

TABLE I. SIOP (Stockholm) Working Classification of Renal Tumors of Childhood (1994)

I. Low risk tumours (favourable)
- Cystic partially differentiated nephroblastoma
- Nephroblastoma with fibroadenomatous-like structures
- Nephroblastoma of highly differentiated epithelial type
- Nephroblastoma—completely necrotic (after preoperative chemotherapy)
- *Mesoblastic nephroma*

II. Intermediate risk tumours (standard)
- Nonanaplastic nephroblastoma with its variants
- Nephroblastoma—necrotic but some features left (<10%)

III. High risk tumours (unfavourable)
- Nephroblastoma with anaplasia
- *Clear cell sarcoma of the kidney*
- *Rhabdoid tumour of the kidney*

IV. Other tumours or lesions
- *Cystic nephroma*
- *Renal cell carcinoma (all variants)*
- *Transitional cell carcinoma*
- *Neuroepithelial tumours:*
  renal neuroblastoma
  renal P.N.E.T.
  renal carcinoid
- *Miscellaneous sarcomas*
- *Renal lymphoma*
- *Angiomyolipoma*
- *Adenomas*
- *Other tumours and lesions*
- *Metastases from other sites*

V. Addendum
Presence/absence of nephrogenic rests should be clearly stated.